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Treatment of Spinal Cord Tumors in Children

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Background. A retrospective review was carried out of nine children under 17 years of age with a diagnosis of intramedullary tumor seen during the period 1989–1995. Six had astrocytomas; one each had an ependymoma, a PNET, and a choroid plexus papilloma. Five patients had back pain, 3 others had mild pareses and the ninth had incapacitating defects. Seven of the 9 were treated by subtotal extirpation of the lesion, and biopsy alone was performed in the other two. All tumors were low grade (grade I or II) and therefore radiation therapy (RT) was performed as the only postoperative treatment in 8 of the 9 children.

Results. In February 1996, seven (77.8%) children were alive and two (22.2%) died of recurrent tumor (7 months and 5 years after diagnosis, respectively). Median follow-up was 3 years 4 months (range: 1 year 6 months to 7 years 3 months).

Conclusion. Surgical removal of intraspinal tumors provides the best hope of control, but spinal column deformity after laminectomy and irradiation is a serious long-term problem in children. Orthopedic supervision for the prevention of these deformities; e.g., by external immobilization, is mandatory. Med. Pediatr. Oncol. 29:293–295, 1997.

Key words: spinal cord tumors; pediatric oncology; iatrogenic deformities

INTRODUCTION

Intramedullary tumors account for 4% to 6% of childhood central nervous system (CNS) tumors and occur sporadically throughout childhood, with a mean age of 10 years. [1,2]. The most frequent histologies are astrocytoma and ependymoma (80%–90%) and the lesions are frequently situated in the thoracic region. Intramedullary astrocytomas and ependymomas are well differentiated, low-grade tumors. High-grade features occur in 11% of astrocytomas and less frequently in ependymomas. Our experience with 9 such patients treated in a single institution is presented here.

MATERIALS AND METHODS

Nine cases of intramedullary tumors in children under 17 years of age (3 males and 6 females) are reported. All patients were treated in the Department of Pediatric Oncology, Prague between 1989 and 1995. The age range was 1 year to 16 years 11 months. Astrocytomas were the most common histologic type, accounting for 6 of the 9 cases. There was one each of primitive neuroectodermal tumor (PNET), ependymoma (grade I), and choroid plexus papilloma (grade I). The first symptoms and signs in our 9 children were pain (55.6%), paresis (33.3%), and headache and vomiting (11.1%). Diagnosis was made by CT, MRI, and histopathologic investigation. The anatomical location of the tumors revealed a predilection for the thoracic region [3], while there were 2 each in the cervical and the conus region of the cord (Table 1).

TREATMENT

Either laminectomy with subtotal extirpation of the lesion or biopsy was performed in all cases. Seven patients (77.8%) had a subtotal resection and 2 (22.2%) had biopsy only. There were no recorded cases of postoperative meningitis. After surgery, 8 patients received local irradiation with tumor doses ranging from 30–44 Gy without complications. One girl did not receive radiotherapy because of parental disagreement. She had 3 operations between May 1992 and March 1993 and is doing well.

RESULTS

Treatment outcomes are summarized in Table 1. Median follow-up was 3 years 4 months, with a range of 1 year 6 months to 7 years 3 months. Two of the 9 children improved immediately after surgery and three following radiotherapy. Two of 8 patients treated by surgery and irradiation died because of local recurrence 7 months and 5 years post surgery. All patients received orthopedic supervision postoperatively, and all survivors have required body braces. In June 1996 six (66.7%) children are in remission (CR), two (22.2%) died and the girl with

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TABLE I. Characteristic of Patients and Outcome

Patient number	Sex	Diagnosis	Localiz.	Surgery	Radioth. (Gy)	State
1	M	Astr.	Th 10–L2	2 × P	44	died
2	F	Astr.	C 3	3 × P	0	alive
3	F	Astr.	C 1–7	2 × P	44	alive
4	F	Astr.	Th 2–12	1 × P	36	died
5	M	Astr.	Th 4–12	1 × P	40	alive
6	F	PNET	C 3–L 2	1 × B	30	alive (PD)
7	F	Epend.	L 2–3	1 × P	40	alive
8	F	Astr.	C 3–Th 5	1 × B	40	alive
9	M	Papil.	Th 2–9	1 × P	40	alive

M—male, F—female, Astr.—astrocytoma, Epend.—Ependymoma, Papil.—Choroid plexus papilloma, PNET—primitive neuroectodermal tumor, C—cervical, Th—thoracic, L—lumbar, P—partial extirpation, B—biopsy, PD—progressive disease.

a PNET (11.1%) has progression of tumor. She is paraplegic with sphincter inadequacy.

DISCUSSION

Recent studies indicate that complete or gross subtotal resections of intramedullary spinal tumors is possible in most patients using new techniques such as the ultrasonic aspirator, and laser beams [4]. Some authors suggest that radical resection of intramedullary tumors in children may be associated with improvement in survival time [4–7] Brumberg et al [2]. suggest combinations of intraoperative sonography and preoperative magnetic resonance imaging to localize the lesion precisely and for establishing the extent of laminectomy and myelotomy. These techniques allow a more specific surgical approach and thus a more complete resection since most ependymomas and low-grade gliomas can be radically resected [8]. Operative mortality is low [4], and none of our patients died of postoperative complications.

Cristante et al. investigated the functional outcome in 69 patients after surgery for intramedullary tumors [8]. Of these with tumors affecting the upper extremities, 17.1% were improved, 55.5% were unchanged, and 31.5% were worse; and when there were lower extremity signs, 22.4% were improved 51.5% were unchanged, and 29.4% were worse. We found an improvement in functional outcome in 5 of our 9 patients (55.6%); 4 were unchanged.

Postoperative spinal deformities are often present and orthopedic follow-up is important [1,4,9–11]. The most frequent deformities in children with intramedullary tumors is scoliosis [9,10,12] affecting especially the thoracic region [5]. In 25% to 40% of children, the development or progression of such deformity occurs within a mean of 3 years [4]. As others have also reported, we found no correlation between the incidence of deformities and the number of laminae removed [12].

The advantage of radiation therapy in the treatment of the intramedullary tumors in children remains [1,3,9, 10,13–15]. Radiotherapy is thought to be unnecessary in radically resected ''low-grade'' tumors, whereas irradiation may be more important following subtotal removal [1,9,10,16,17].

Concerning ''high-grade'' astrocytomas, radiation therapy is recommended but with uncertain outcome [1,3,18]. However, it has been found to increase disease-free survival time in children with subtotally resected ependymomas [5,19,20]. A dose of 45Gy or even 50Gy is recommended to the area of the involvement [4]. We have only one patient with ependymoma and she is doing very well, without neurological complications after subtotal tumor removal and 40Gy. Data concerning irradiation of primary intramedullary PNET are rare. Our experience with one patient shows that treatment with vincristine, carboplatin, cyclophosphamide, etoposide, and radiotherapy (30 Gy) may not be sufficient for this tumor. She is paraparetic, and a recent MR shows new progression of the tumor.

High dose radiation therapy has unfavorable side effects on the spinal cord and growing spine. Radiation myelopathy can either be transient or a progressive process leading to complete transverse myelopathy [13,21]. Our experience to date suggests that a dose of 40Gy for low-grade tumors is sufficient, but longer follow-up of our patients is needed before definitive conclusions can be reached.

Neurofibromatosis (NF) type 2 is associated with various peripheral nervous system tumors including intramedullary spinal cord ependymomas [22]. Therefore, the follow-up of patients suffering from NF is important. In one patient reported here who suffered from NF there was a rapid progression of an astrocytoma despite irradiation that led to his death 7 months after operation.

CONCLUSION

The most effective treatment of intramedullary tumors is radical surgery. This type of surgery is now possible

more often since the advent of modern imaging techniques; i.e., CT, MRI, and intraoperative sonography. Radiation therapy should be reserved for ''high-grade'' tumors (astrocytoma, ependymoma) or for recurrence of subtotally removed ''high-grade'' lesions. Irradiation after partial removal of ''low-grade'' tumors can provide a better prognosis for these types of tumors. All children must be monitored by an orthopedist for proper management of iatrogenic deformities.

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